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Fibrous Dysplasia of the Spine

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Fibrous dysplasia can involve any part of the skeleton. However, the spine is a relatively uncommon site, especially for the monostotic form. We have reviewed five cases of fibrous dysplasia of the spine; two were monostotic and three were polyostotic. There was no predilection for any particular level of the spine, and both the posterior elements and vertebral bodies were involved. The typical radiologic appearance was a well defined, expanding lesion with variable density, similar to the lesions in the appendicular skeleton. Clinically, spinal lesions were often symptomatic due to nerve root encroachment or secondary arthritis, even in the absence of a pathologic fracture.

Fibrous dysplasia is considered to be a developmental anomaly characterized by the failure of differentiation of bone-forming mesenchyme.[4] Although any bone can be involved, the spine is rarely affected, especially in the monostotic form.[5] The radiographic appearance is usually typical. However, it frequently presents a diagnostic dilemma. In this report, we studied five histologically proven cases of fibrous dysplasia of the spine.

Five cases of fibrous dysplasia of the spine, including the sacrum, were collected between 1982 and 1988. In all these cases, the diagnosis was confirmed histologically. Medical records, and radiologic and pathologic studies were reviewed.

Patients' ages ranged from 19 to 38 years (mean, 30 years); there was one man and four women. One patient was asymptomatic, an incidental finding on radiographs (Figure 1). The other four were symptomatic and presented with low-back pain and/or hip pain. The monostotic cases (two of five) involved the lateral mass of C1 (Figure 1) and the body of L3. One of the three polyostotic cases presented with involvement of adjacent articular facets of the L3 and L4 (Figure 2). The other two cases involved the sacrum, as well as bones outside the spine—including the sternum and humerus. Overall spinal involvement included the vertebral body (two cases), facet (two cases, three lesions), and the sacral wing (two cases). Four of the cases were predominantly lytic, and one was mainly blastic (Figure 2). All five lesions (four cases) showed expansion. One case showed destructive changes in the facets from arthritis (Figure 2).

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Histology

Histologically, all of the lesions were similar. The spindle cell component was cytologically benign, moderately cellular, and the cells were arranged in short intersecting fascicles. Present within the spindle cell regions were disorganized and irregularly shaped bony trabeculae (Figure 3). The trabecular bone was woven in architecture and lacked prominent osteoblastic rimming. The histologic features were characteristic of fibrous dysplasia. In one case involving the sacrum, the fibrous dysplasia showed evidence of cystic degeneration.

Discussion

Fibrous dysplasia is a multisystem disorder that may involve the central nervous system, endocrine organs, skin, and the skeleton. Skeletal involvement can be monostotic (70–80%) or polyostotic (20–30%).[1] In the monostotic form, long tubular bones are often involved. On the other hand, the axial skeleton is more often involved in the polyostotic form. Involvement of the spine is rare, especially in the monostotic form. Resnik and Lininger reviewed five reported cases of monostotic form of the spine and added one case.[5] Based on reported cases[5] and our series, there is no apparent predilection for

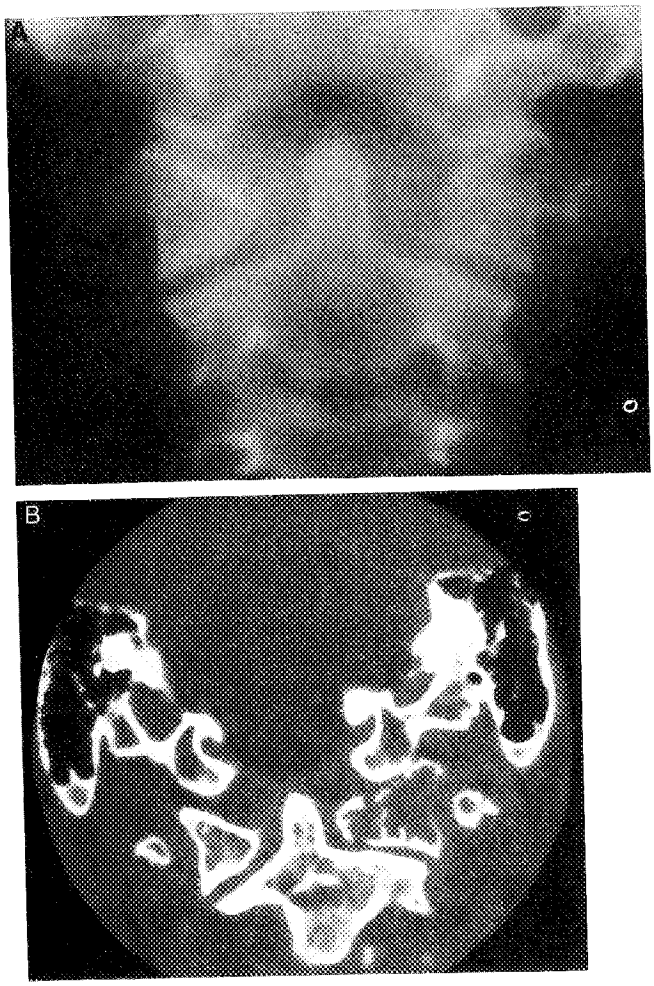

Figure 1. Monostotic fibrous dysplasia of C1 (19-year-old man). **A,** Anteroposterior tomogram of C1 shows an expanding lytic lesion with coarse trabeculation in the left lateral mass. **B,** Computed tomography scan of C1 shows a well defined lytic lesion with coarse trabeculation and a pathologic fracture of the left lateral mass.

any particular portion of the spine. The posterior elements as well as the vertebral bodies are affected.

Well defined, purely lytic lesions with homogenous density and expansion of the cortex were characteristic findings in our series. Blastic change was seen in one case, and coarse trabeculation simulating vertebral hemangioma was seen in another. There was no significant difference in the radiologic appearance when compared to fibrous dysplasia in other regions of the skeleton.[2] With lytic lesions, hemangioma, giant cell tumor, and aneurysmal bone cyst should be considered in the differential diagnosis. Computed tomography and magnetic resonance imaging are useful to rule out an aggressive process with cortical destruction or soft tissue extension. If blastic, Paget's disease and osteoblastoma should be included in the differential diagnosis. Because of the relatively young age, multiple myeloma and metastatic carcinomas are considered less likely possibilities.

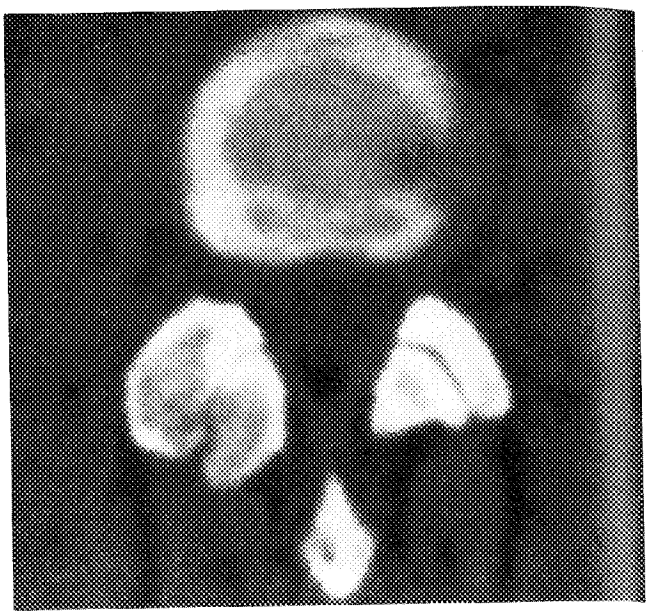

Figure 2. Polyostotic fibrous dysplasia of the adjacent articular facets of L3 and L4 (38-year-old woman). No other part of the skeleton was involved. Computed tomography scan at L3–L4 level shows the expansion of the right facets of L3 and L4 with bony erosion on both sides of the facet joint. Density is homogenous, and the characteristic ground glass appearance is seen.

Fibrous dysplasia of tubular bones usually becomes symptomatic only if a pathologic fracture occurs or if the deformity of the weight-bearing bone is severe enough to cause limping.[3] However, the lesions involving the spine are often symptomatic due to nerve root encroachment, even in the absence of a pathologic fracture. The destructive arthritis of the facet joint in one of our cases was rather unusual.

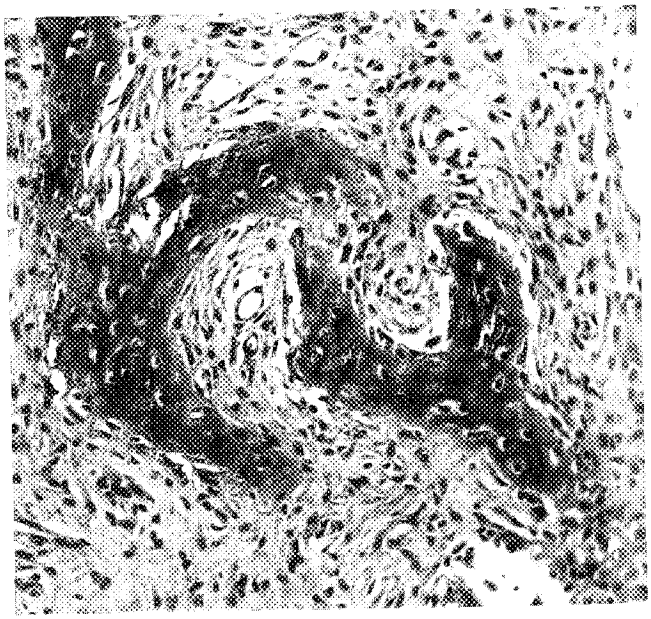

Figure 3. Histologically dysplastic curvilinear trabeculae of woven bone within a spindle cell fibrous background are seen. Rimming osteoblasts are inconspicuous (H&E; original magnification ×200).

In the polyostotic form with typical lesions in the appendicular skeleton, diagnosis is not difficult. In monostotic fibrous dysplasia, biopsy may be indicated because metastatic disease or multiple myeloma may simulate a benign, nonaggressive process.

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Intramedullary Lipoma

Diagnosis and Treatment

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Spinal cord tumors are uncommon. After a 10-year observation period, Guomundsson reported an incidence of 1.1% spinal tumors of all types per 100,000 population.[5] Most of these tumors are benign extramedullary lesions, with intramedullary tumors comprising approximately 16%.[7] Ependymomas and astrocytomas are the most common intramedullary lesions by far, with lipomas comprising 2% in a Mayo Clinic review of 1,322 spinal cord tumors.[9] Ehni and Lore reported the incidence of lipomas as 1% of primary spinal cord tumors, with 40% of them being extradural and often associated with various degrees of dysraphism, particularly spina bifida occulta.[4]

Presented here is an exceedingly rare occurrence of an intramedullary lipoma that heralded symptoms in the sixth decade and was not associated with any roentgenographic abnormality. Since most neurosurgeons would not encounter this lesion in their lifetime, I was prompted to review its diagnosis and treatment.

Case Report

A 67-year-old Japanese woman presented with slowly progressive lower extremity weakness. The right limb was more affected than the left limb. The degree of weakness vacillated from time to time as well. On a recent extended vacation, the leg weakness was complicated by recurrent limb numbness and sclerotomal pain, but no sphincter dysfunction.

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Examination demonstrated bilateral lower extremity hyperreflexia, extensor toe signs on the left, mild bilateral paraparesis, and a partial sensory level to pin-prick to T11 on both sides. Position sense was affected on the right, and the saddle dermatomes appeared partially implicated bilaterally as well, although there was no sphincter dysfunction.

Investigation. Thoracolumbar roentgenograms revealed advanced lumbar degenerative disease. Omnipaque myelography completely missed the lesion (Figure 1), but the subsequent

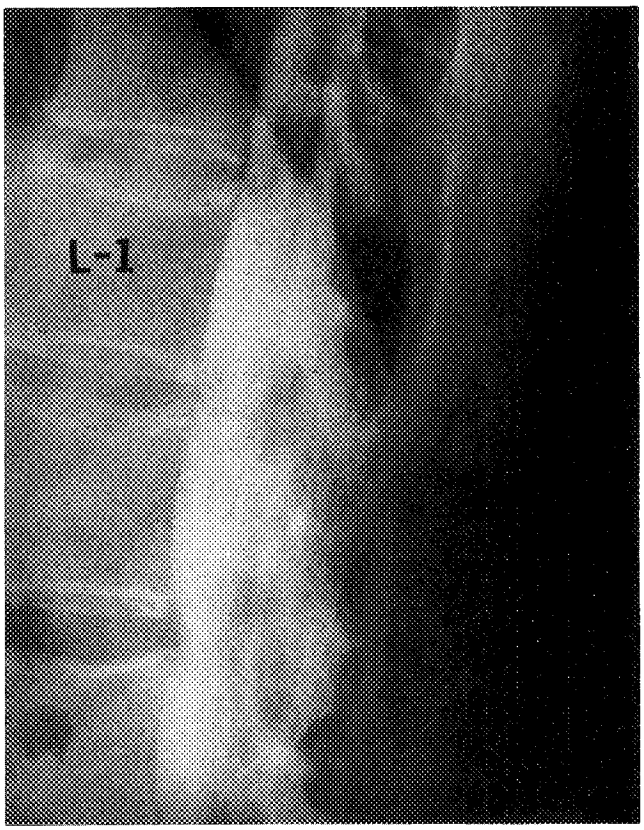

Figure 1. Myelography was nondiagnostic.